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Unusual presentation of fibrous dysplasia

A case of fibrous dysplasia is described in which the unusual site and manner of presentation delayed diagnosis. Fibrous dysplasia occurs primarily in children. The initial symptom of facial bone involvement is usually painless facial asymmetry. The child we report presented at age 12 with her third episode of left-sided acute dacryocystitis and no bony facial asymmetry. She was found to have a mucosa-covered mass originating on the left lateral nasal wall and almost entirely obstructing the nasal airway.

A CT scan revealed a large calcified mass involving the inferior turbinate. Biopsy suggested fibrous dysplasia. The patient subsequently underwent resection of all involved bone via a sublabial degloving approach (chosen to avoid external facial scarring). At one year after surgery, the patient has an excellent cosmetic result, a patent nasal airway, and no epiphora.

Facial asymmetry in late childhood or early adulthood is the usual presentation of fibrous dysplasia of the facial bones. It is more common in females, and up to 25% of these patients may have a second site of skeletal involvement (1). Up to 10% of cases of monostotic fibrous dysplasia involve the craniofacial bones (2).

The etiology of fibrous dysplasia remains unknown. It was thought at one time that the disease occurred in a site of previous trauma, but more recent studies have failed to confirm this correlation (3).

Diagnosis of fibrous dysplasia is made by biopsy. The tissue shows a fibrous connective tissue matrix with scattered bony or osteoid trabeculae (4). Radiographs of these lesions may show lucent or sclerotic areas, depending on the ratio of connective tissue to bone (5).

Treatment of fibrous dysplasia is usually conservative, since it is a benign process with less than 0.5% incidence of malignant transformation (6). The disease often becomes inactive at the time of puberty, but this is variable and, occasionally, no arrest occurs (7). Small asymptomatic lesions require only periodic observation. Cosmetic deformity is the most common indication for surgery, and partial removal with curettage can be performed for these cases (5,8). Occasionally the lesions cause dysfunction of adjacent structures, and more aggressive surgical removal is necessary. Even with apparently complete removal there is a 10% to 20% incidence of recurrence.

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A case is presented here in which the lesion's site and manner of presentation were unusual, thereby delaying the diagnosis.

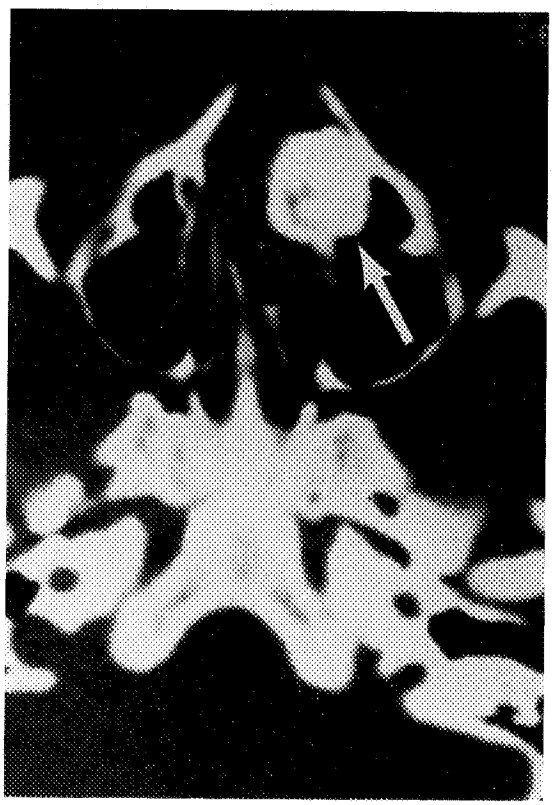
Case report

A 12-year-old girl presented with her third episode of left-sided dacryocystitis in three years. She had no epiphora between episodes and no nasal obstruction. She was referred to the otolaryngology service when a CT scan showed an intranasal mass.

Physical examination revealed an erythematous fluctuant mass at the left medial canthus. Intranasally, the septum was slightly deviated, and there was a large mucosa-covered mass on the left lateral nasal wall. Vision was normal, as was the remainder of the head and neck examination. The CT scan (Fig 1) showed a calcified mass adjacent to a walled-off abscess in the area of the left medial canthus.

Her abscess was treated by incision and drainage and appropriate intravenous antibiotics (chloramphenicol and gentamicin). Biopsy of the intranasal mass revealed a proliferation of fibrous tissue with scattered trabeculae of immature bone, consistent with the diagnosis of fibrous dysplasia (Fig 2).

Several weeks after the infection had resolved, the patient underwent a left partial medial maxillectomy through a sublabial approach to the midface



1. CT scan shows dense mass in the medial maxilla and inferior turbinate area.

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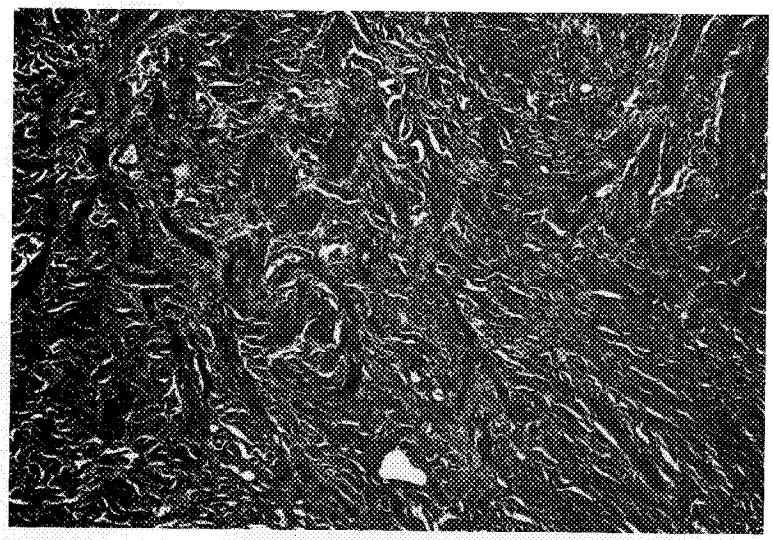
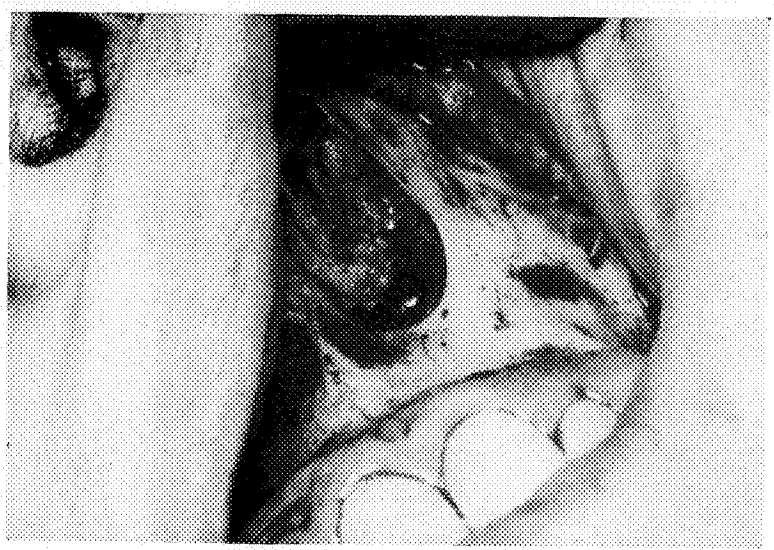
(9) (Fig 3). Fibrous dysplasia involved the anterior two thirds of the inferior turbinate and the lateral nasal wall. The involved bone was soft, very vascular, and blended indistinctly with normal bone. A dacryocystorhinostomy was performed at the same time by the referring ophthalmologist. The patient's postoperative course was uneventful. One year after surgery the patient had a patent nasal airway with no epiphora.

2. Photomicrograph showing trabeculae of immature bone within fibrous tissue (sometimes referred to as a Chinese writing *pattern).*





3. Intraoperative photo showing degloving of soft tissue of midface. The enlarged inferior turbinate with soft, coarsely-textured bone is visible.

Discussion

In this case the diagnosis of fibrous dysplasia of the medial maxilla and inferior turbinate was delayed by an unusual presentation, recurrent acute dacryocystitis. Dacryocystitis is most often secondary to fibrotic obstruction within the lacrimal drainage system. This is frequently idiopathic. Tumors and other causes of external obstruction are relatively uncommon (10,11). In this case the patient probably had chronic dacryocystitis from constant impingement on the lacrimal duct by the fibrous dysplasia mass. It is interesting that despite a large intranasal mass, she had no nasal symptoms or facial asymmetry. Her third episode of acute dacryocystitis prompted her ophthalmologist to order a CT scan which led to the diagnosis of an intranasal mass. An adequate intranasal examination earlier in the patient's course may have led to an earlier diagnosis.

To our knowledge, involvement of the inferior turbinate by fibrous dysplasia in the manner described in this case has not been previously reported. The sublabial degloving approach to the midface provided excellent exposure and avoided the need for a lateral rhinotomy. The patient has a good cosmetic result with no nasal deformity or large facial scar.

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